AB1018

TREATMENT RESPONSE OF HYDRONEPHROSIS ASSOCIATED WITH IDIOPATHIC RETROPERITONEAL FIBROSIS, FOCUSING ON RATIO OF IGG4/IGG3 SERUM CONCENTRATION

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Background: Hydronephrosis, a common complication of idiopathic retroperitoneal fibrosis (iRPF), may lead to poor renal outcomes unless it is resolved. Pathological confirmation can help to identify the aetiology of the disease and determine the treatment strategy. But, in most cases, it is difficult to obtain sufficient tissue due to the location of fibrosis. In a recent study, parts of iRPF are correlated with IgG4-related disease characterised by elevated serum IgG4 levels (>135 mg/dL). Normal serum IgG3 level (21–176 mg/dL) has been known to be higher than normal serum IgG4 level (4–86 mg/dL). The reverse IgG4/IgG3 ratio has been suggested to be an IgG4-related disease component that distinguishes it from primary sclerosing cholangitis [1]. However, the ratio of IgG3 and IgG4 may be reversed in iRPF patients with hydronephrosis.

Objectives: We aimed to investigate the ratio of IgG subclasses as a predictive factor associated with treatment response of hydronephrosis in patients with iRPF.

Methods: We retrospectively recruited 19 iRPF patients with hydronephrosis who evaluated serum IgG subclasses in a tertiary hospital between 2004 and 2019. Hydronephrosis was evaluated on the basis of imaging findings. Medications and clinical and laboratory findings, including IgG subclasses, were reviewed following the diagnosis of hydronephrosis. Hydronephrosis improvement on subsequent images was evaluated to assess treatment response. Categorical data were compared using chi-square or Fisher's exact test. Continuous variables were compared using Mann-Whitney U test.

Results: At baseline, median serum IgG3 and IgG4 levels were 64 (IQR 37–82) mg/dL and 71 (IQR 40–171) mg/dL. Five patients had serum IgG4 levels > 135 mg/dL and 11 patients had the reverse serum level of IgG4/IgG3. On subsequent images (median follow-up at 3.2 [IQR 1.7–4.0] months), 11 patients showed hydronephrosis improvement. The proportions of positive ratio of serum IgG4/IgG3 (81.8% vs. 25%, $p = 0.024$), periaortic involvement (81.8% vs. 25%, $p = 0.024$) and high-dose glucocorticoid treatment (45.5% vs. 0%, $p = 0.045$) were significantly higher in patients with improvement than in those without improvement (Table 1). Interestingly, even in cases with normal serum IgG4 levels, patients with improvement showed a higher serum IgG4/IgG3 ratio than in those without improvement (median 1.5 vs. 0.7, $p = 0.038$).

Table 1. Clinical characteristics and treatment according to the short-term outcome of hydronephrosis

	Improvement (n = 11)	No improvement (n = 8)	p-value
Time to subsequent imaging (months) ^a	2.2 (1.3–4.2)	3.2 (2.0–3.8)	0.778
Males (n, %)	8 (72.7%)	7 (87.5%)	0.603
Periaortic involvement (n, %)	9 (81.8%)	2 (25%)	0.024
Impaired renal function (n, %) ^b	5 (45.5%)	2 (25%)	0.633
Serum IgG4 (mg/dL) ^a	114 (59–172)	43 (35–109)	0.152
Elevated serum IgG4 (n, %)	4 (36.4%)	1 (12.5%)	0.338
Serum IgG4/IgG3 ratio ^a	2.1 (1.2–4.9)	0.8 (0.4–1.0)	0.041
Positive ratio of serum IgG4/IgG3 (n, %)	9 (81.8%)	2 (25%)	0.024
Medical treatment (n, %)	7 (63.6%)	1 (12.5%)	0.059
High-dose glucocorticoid treatment (n, %)	5 (45.5%)	0 (0%)	0.045
Surgical intervention (n, %)	8 (72.7%)	4 (50%)	0.377

^a Values are median and interquartile range (25th–75th percentile)

^b Defined as serum creatinine level > 1.24 mg/dL

Conclusion: The reverse ratio of serum IgG4/IgG3 was associated with hydronephrosis treatment response, thus suggesting favourable responses to high-dose corticosteroid.

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AB1019

THERAPEUTIC PROFILE OF PATIENTS WITH VOGT KOYANAGI HARADA DISEASE FOLLOWED AT THE HOSPITAL DE CLÍNICAS. SAN LORENZO PARAGUAY.

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Background: Vogt-Koyanagi-Harada Disease (VKHD) is an autoimmune eye disease that causes chronic vision complications in young people. The treatment is based on corticosteroids and immunosuppressants, to which biological drugs are currently added. The visual prognosis depends on the stage in which it is diagnosed and the effective and timely treatment.^{1,2}

Objectives: Describe the evolution and the therapeutic profile of patients with VKHD who go to the Rheumatology Department of the Hospital de Clínicas. San Lorenzo Paraguay until November 2019

Methods: Descriptive cross-sectional study conducted at the Hospital de Clínicas, San Lorenzo Paraguay, where clinical histories of patients with this disease and who have regular follow-up were reviewed.

Results: 21 patients were included, 17 (81%) women, average age of 35 years. The diagnosis and treatment were made 15 (71%) in acute phase, 4 (19%) convalescent phase and 2 (10%) in chronic phase.

Corticosteroids: All received corticosteroids at diagnosis. Methylprednisolone IV 1g /day for 3 consecutive days in 8 (44%) cases and for 5 days in 10 (55.5%) cases. The first month everyone received prednisone orally at 1mg / kg / d on average (60–75 mg/d), at 3 months the average dose of prednisone was 37 mg / d, at 6 months of 23 mg/d and at 12 months 10 mg / d months.

Immunosuppressive treatment: They received 20 (95%) patients. Induction with monthly intravenous cyclophosphamide was performed in 8 (40%) cases and with Azathioprine in 12 (60%) cases. Of the patients treated with Azathioprine initially, in 5 (23%) patients it was necessary to switch to Cyclophosphamide IV due to failure or intolerance. The maintenance treatment was azathioprine in 13 (62%) of the patients, MTX in 4 (19%) and the association of both drugs was indicated in 3 (14%) patients. One case was refractory and received infliximab and subsequently Rituximab, which resulted in remission.

The best final Visual Acuity (AV) was recorded with the Snellen chart in 40 eyes (20 patients), 27 (67.5%) patients with good vision (20/20 to 20/50), 8 (20%) patients with decreased VA (20 / 50-20 / 200), and 5 (12.5%) with legal blindness (<20/200) and of these only 3 patients had unilateral involvement and one patient with bilateral blindness. Sequelae / complications: 6 (28.5%) patients presented cataracts and finally 3 (14%) glaucoma, 5 (24%) anterior / posterior synechiae.

Conclusion: The majority of patients in this series presented a good vision when the inflammatory process was resolved (AV 20 / 20-20 / 50), this probably related to early diagnosis and initial aggressive treatment with intravenous corticosteroid pulses and its association with immunosuppressants of entry as mentioned in the literature. The worst vision, (visual acuity <20/200) occurred in 5 of 40 eyes, corresponding to 4 patients, two of them diagnosed and treated in the chronic stage. Timely treatment improves visual prognosis.

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AB1020

APPLICATION OF THE 2019 ACR/EULAR CLASSIFICATION CRITERIA CONTRIBUTES TO THE DIAGNOSIS OF IGG4-RELATED DISEASE

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Background: IgG4-related disease (IgG4-RD) is a rare and systemic disease characterized by swelling or masses in the involved organs, elevated serum IgG4, marked IgG4+ plasma cells infiltration and fibrosis. However, lacking of pathological data limited the definite diagnosis of IgG4-RD. As results of high-quality clinical and epidemiological investigations collected, new classification criteria for

IgG4-RD were approved by ACR and EULAR in 2019. Whether this new criteria improve the diagnosis efficiency needs to be validated in clinical practice.

Objectives: To apply the 2019 ACR/EULAR classification criteria for the diagnosis of IgG4-RD in previously suspected patients and explore the clinical characteristics of patients with IgG4-RD according to the new classification criteria.

Methods: Patients suspected of having IgG4-RD due to elevated serum IgG4 and swelling or masses in single or multiple organs were recruited in Sun Yat-Sen Memorial Hospital, Sun Yat-Sen University from May 2013 and November 2019. Demographic and clinical data were collected. The diagnosis was reevaluated with the 2011 comprehensive diagnostic criteria (CDC) for IgG4-RD and the 2019 ACR/EULAR classification criteria for IgG4-RD, respectively.

Results:

- There were 68 patients recruited and 59(86.8%) of them had elevated serum IgG4 ($\geq 135\text{mg/dl}$) and 53(77.9%) patients showed swelling or masses in single or multiple organs. Most patients first visit general surgery (17.6%), gastroenterology (16.2%), respiratory medicine (16.2%) and rheumatology (14.7%).
- According to the 2011 CDC for IgG4-RD, 4(5.9%) patients were definite IgG4-RD, 1(1.5%) was probable and 42(61.8%) were possible. According to the 2019 ACR/EULAR criteria, 20(29.4%) patients were diagnosed as IgG4-RD, including the 4 definite patients using the 2011 CDC.
- Among the 20 IgG4-RD patients according to the 2019 ACR/EULAR criteria, 19(95.0%) were male and median age of symptom onset was 62(46–69) years. There were 6(30.0%) patients diagnosed at rheumatology, 5(25.0%) at gastroenterology, 3(15.0%) at general surgery, 2(10.0%) at respiratory medicine and 1(5.0%) at stomatology, endocrinology, orthopedics and urinary surgery, respectively. There were 9(45.0%) patients with bilateral lacrimal or salivary glands involved, 9(45.0%) with pancreas and biliary tree involved, 5(25.0%) with retroperitoneum involved, 2(10.0%) with kidney involved and 1(5.0%) with chest involved.
- The median serum IgG4 of the 20 IgG4-RD patients was 15.40(4.14–55.10) g/L, median serum IgG was 27.9(17.2–50.2)g/L. There were 20.0%(4/20) patients had elevated serum eosinophil and 93.3%(14/15) had elevated serum IgE. There were 60.0%(9/15) patients had elevated C-reactive protein, 85.7%(12/14) had elevated erythrocyte sedimentation rate and 26.7%(4/15) had hypocomplementemia. There were 35.7%(5/14) patients had positive rheumatoid factor and 14.3%(2/14) had positive antinuclear antibodies. There were 6(30.0%) patients didn't received biopsies, 8(40.0%) patients received surgical removal of salivary glands, lacrimal glands or pancreatic masses and 6(30.0%) patients received needle biopsies of the salivary glands or biopsies by bronchoscopy, gastroscopy or enteroscopy.
- Among the patients didn't fulfill the 2011 CDC and the 2019 ACR/EULAR criteria, there were 40 patients had elevated serum IgG4, whose serum IgG4 level were significant lower than those of IgG4-RD patients [3.66(2.39–7.68) g/L vs. 15.40(4.14–55.10)g/L, $P < 0.001$], and percentage of serum IgG4 $\geq 5\times$ upper limit of normal were also lower than those of IgG4-RD patients (27.5% vs. 73.7%, $P = 0.002$).

Conclusion: The 2019 ACR/EULAR criteria can help diagnosing patients with IgG4-RD even lacking the tissue confirmation. Patients with moderately elevated serum IgG4 need more clinical evidence to diagnose IgG4-RD and exclude mimics.

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AB1021

CHARACTERISTICS AND MANAGEMENT OF RHEUMATIC MANIFESTATION UNDER ESTROGEN RECEPTOR-TARGETING CANCER THERAPIES, DATA FROM A PROSPECTIVE REGISTRY

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Background: The knowledge about interdependencies between rheumatic manifestations and malignancies is limited. Further, reliable data on the occurrence of rheumatic symptoms as side effects of specific cancer therapies beyond checkpointinhibitor-induced immune-related adverse events are sparse. In this regard, although arthralgia under estrogen receptor-targeting therapies (aromatase inhibitors and the estrogen receptor modulator tamoxifen) has been frequently reported in oncological clinical trials and case reports, prospective

data including an assessment of rheumatic manifestations by rheumatologists are lacking.

Objectives: To contribute to a better understanding of interdependencies between rheumatic manifestations and cancer/ estrogen blockade and potentially improve the management of both entities, pilot data were analysed.

Methods: Data on characteristics and treatment of rheumatic manifestations and cancer as well as their timely association were systematically, prospectively collected and analysed in the Malheur ('malignancy and rheumatic disease') registry, a long-term, observational study designed to study patients suffering from concomitant rheumatic disease and malignancy and/or premalignant lesions.

Results: We identified 11 patients with rheumatic manifestations under estrogen receptor-targeting therapies (3 anastrozol, 4 letrozol, 8 tamoxifen) as part of breast cancer treatment. In addition to breast cancer one patient had a lymphoma 3 years after and another patient had a non-small cell lung cancer 2 years before breast cancer diagnosis. The patients had different cancer stages (5 IA, 3 IIA, 1 IIB, and 1 IVA). Their mean age at cancer diagnosis was 60.4 ± 11.6 years and all patients are females. The time interval between diagnosis of cancer and onset of systemic/ rheumatic symptoms was 49.5 ± 34.0 months. Of interest, the time interval between onset of rheumatic symptoms and first assessment by a rheumatologist was 16.9 ± 22.3 months. The following systemic and rheumatic symptoms were reported: arthralgia in 10, arthritis in 8 (small joints in 5, large joints in 3 affected), morning stiffness ($>30\text{min}$) in 7, IBP in 1, myalgia in 7, sicca symptoms in 2, fever in 1 (new-onset FMF with heterozygous M694U mutation), class IV glomerulonephritis and polyserositis in 1 (with new-onset SLE) patient(s). Disease burden at baseline was rather high with a mean VAS pain of $65 (\pm 12.9)/100$. Laboratory analyses revealed an increased CRP in 6/11 (55%) with a mean of $10.3 \pm 8.2\text{mg/l}$ (<5). Autoantibody positivity was observed for ANAs in 5/10 (50%, titers ranging from 1:80 to 1:160), anti-dsDNA in 1, rheumatoid factor in only 1/10 (10%) patients, none was anti-CCP positive. Before consulting a rheumatologist, patients were treated with NSAR 3/11 (27%), 10/11 systemic glucocorticoids (91%) with an initial dose of $17.5 \pm 19.5\text{mg}$ and intra-articular glucocorticoids 1/11 (9%). Rheumatological assessment lead to initiation of csDMARDs (3/11 MTX, 1/11 SSZ, 1/11 HCQ, 1/11 AZA (later MMF/ rituximab in the SLE patient) 1/11 colchicine) as corticosteroid-sparing agents with good response in the majority of patients.

Conclusion: Our data demonstrate heterogeneous rheumatic manifestations, partially with severe manifestations beyond arthralgia, so far not reported by oncological studies including follow-up, which might suggest an underreporting. Furthermore, despite close monitoring in tumor aftercare, our data show a considerable delay in referral to a rheumatologist and initiation of suitable treatment. The prospective design of the Malheur registry enables future validation of our pilot data.

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AB1022

PANNICULITIS IN RHEUMATOLOGICAL PRACTICE: THE RESULTS OF LONG-TERM FOLLOW-UP

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Background: Panniculitis is a group of heterogeneous inflammatory diseases that occur with damage to the subcutaneous fat (SCF), musculoskeletal system, and often internal organs

Objectives: to evaluate the course and outcome in patients with panniculitis

Methods: the course of the disease was monitored in 193 patients treated at the V.A. Nasonova Research Institute of Rheumatology for 3-5 years (142 women, 51 men) aged 19 to 77 with the referral diagnosis of erythema nodosum (EN) and